

Fig. 1a Non-Transduced hMSCs in Immuno-competent mouse muscle Day 3

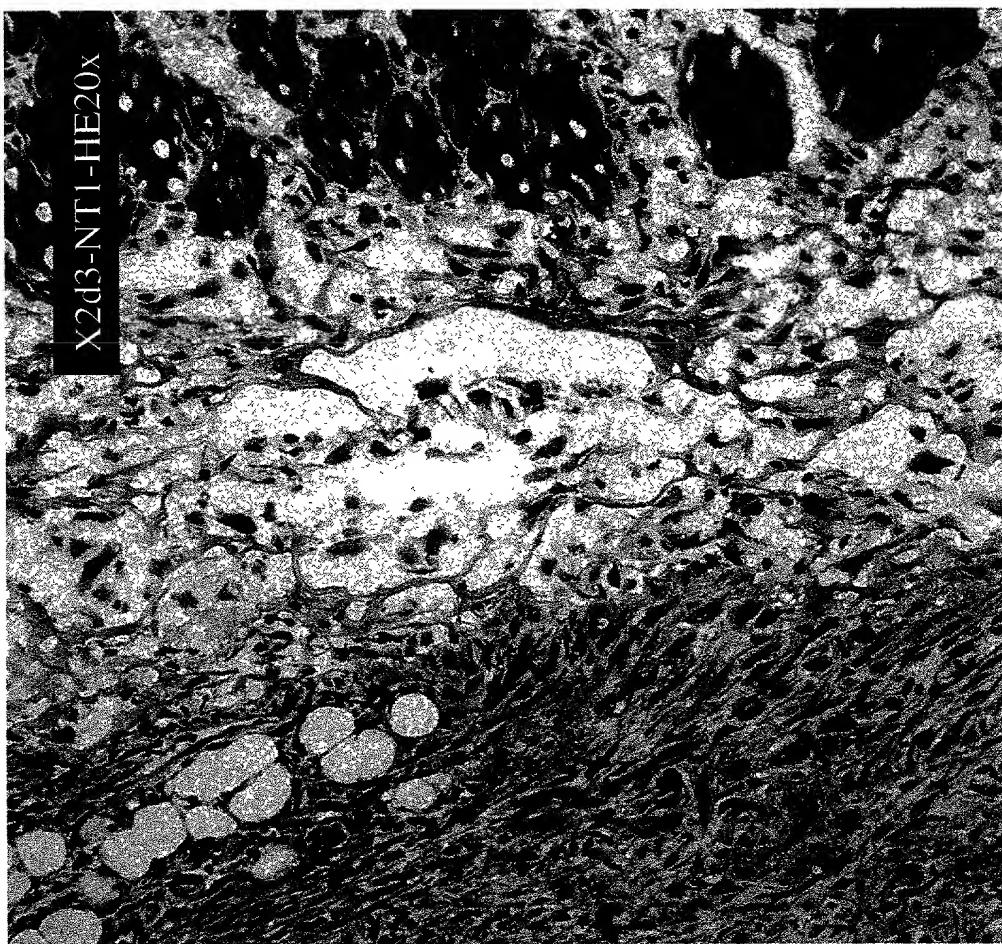
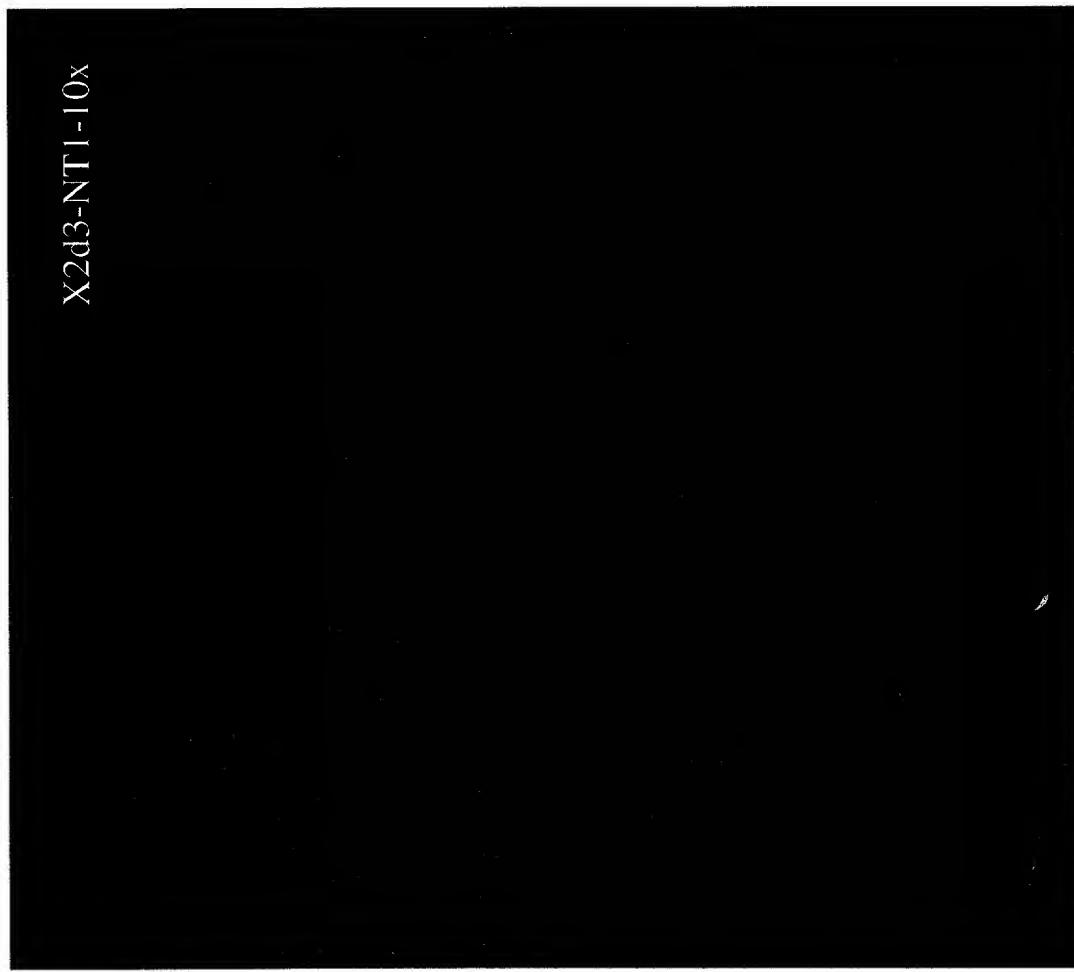
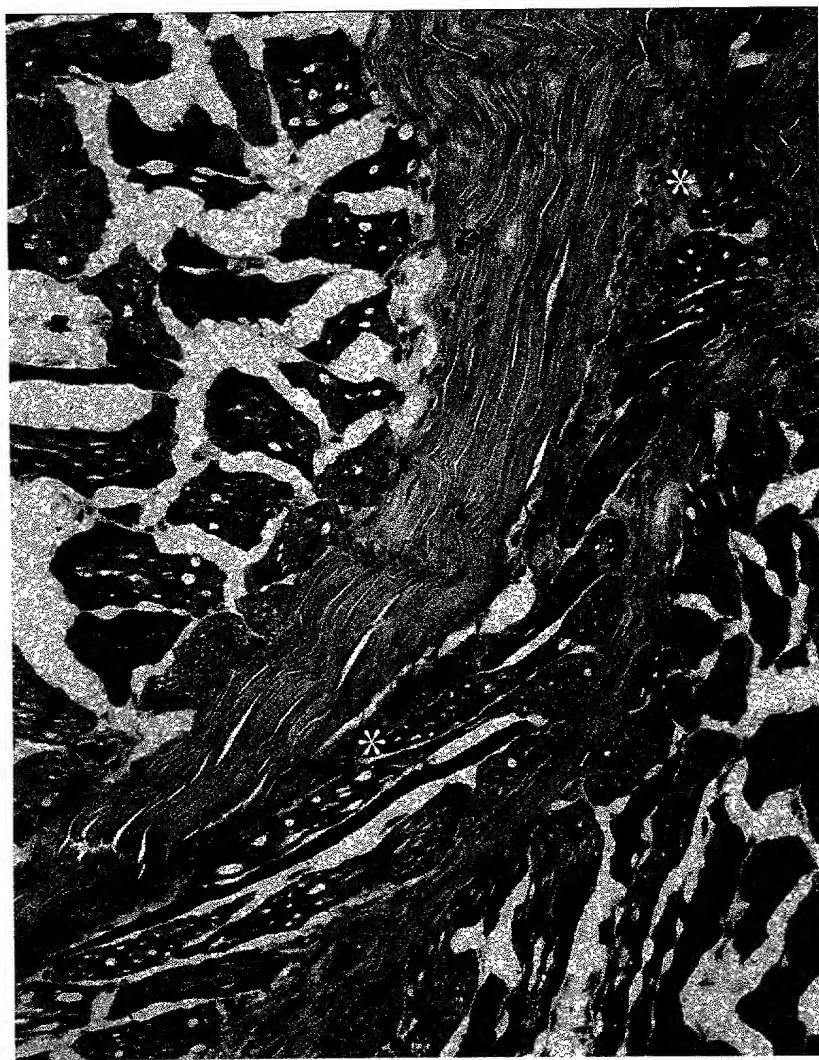


Fig.1b

Human MSCs in Mouse Muscle: Day-7

(H&E)



Dil labelled cells +
Sybr-green staining
of all nuclei

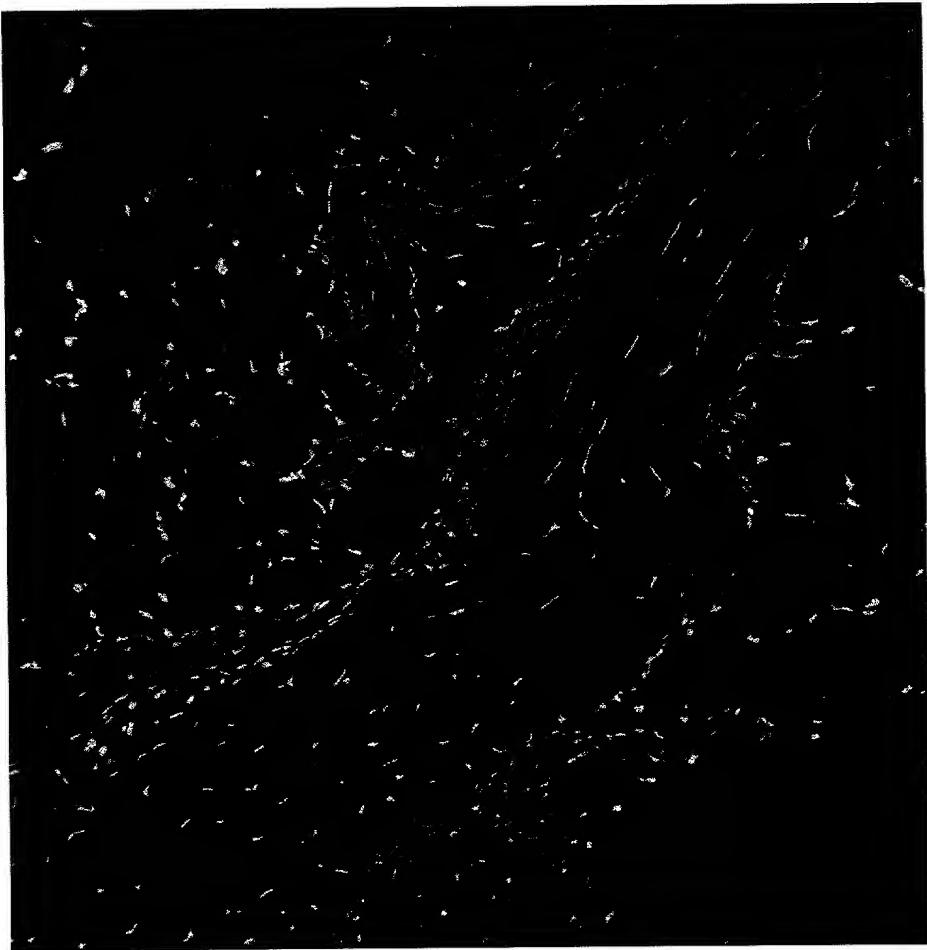


Fig.1c Human MSCs in Mouse Muscle: Day-7

(H&E, 20X)

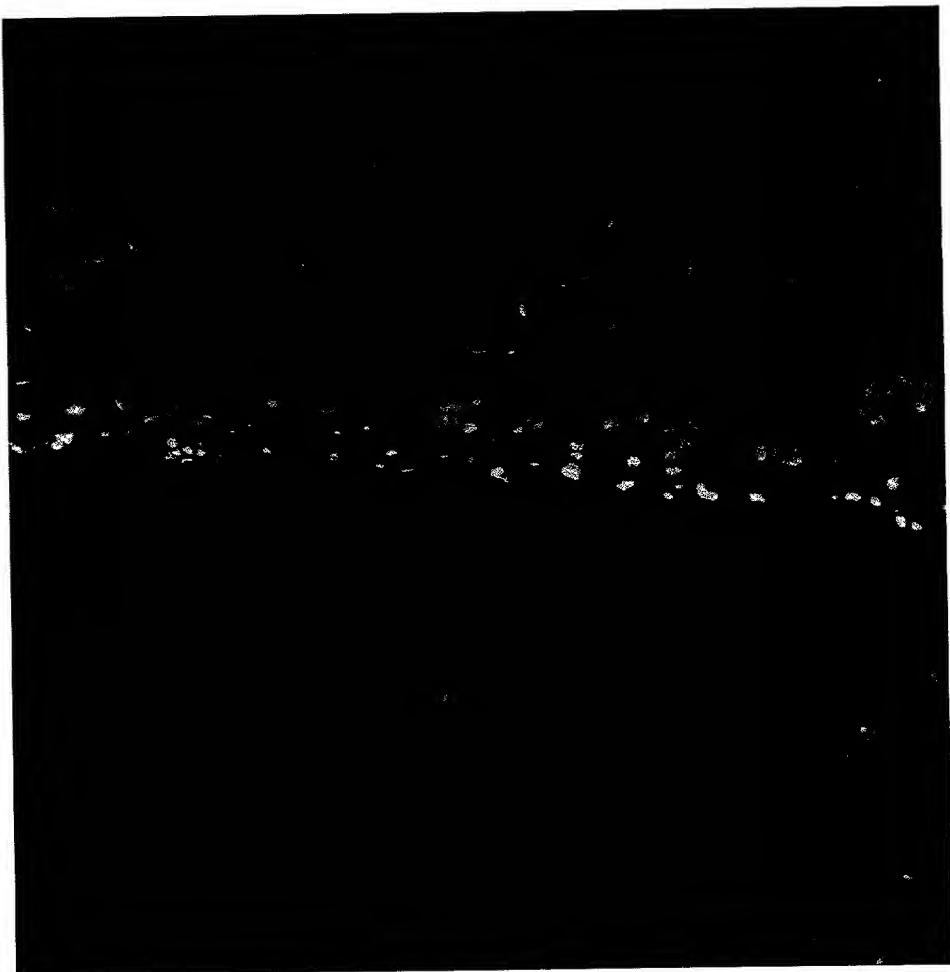
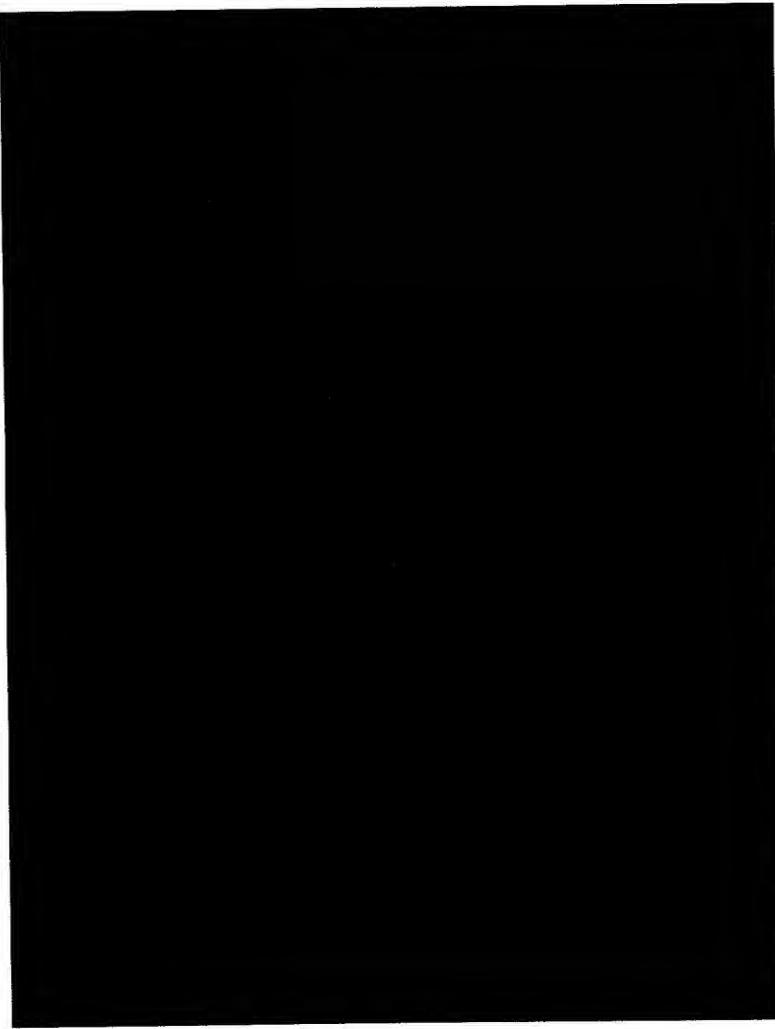


Fig. 1d

Human MSCs in Mouse Muscle: Day 14

Fluorescent Field, 10X



H&E 40X

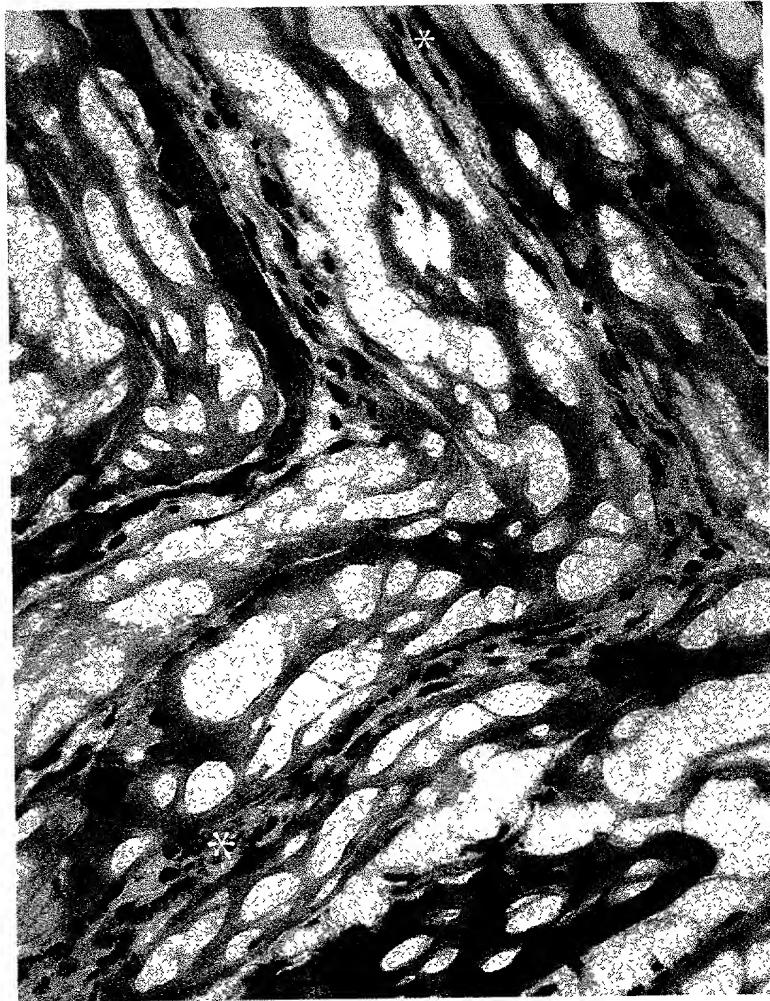


Fig. 1e Dil⁺-Positive α -GalA Transduced hMSCs in

Immuno-competent mouse muscle

d29

d43

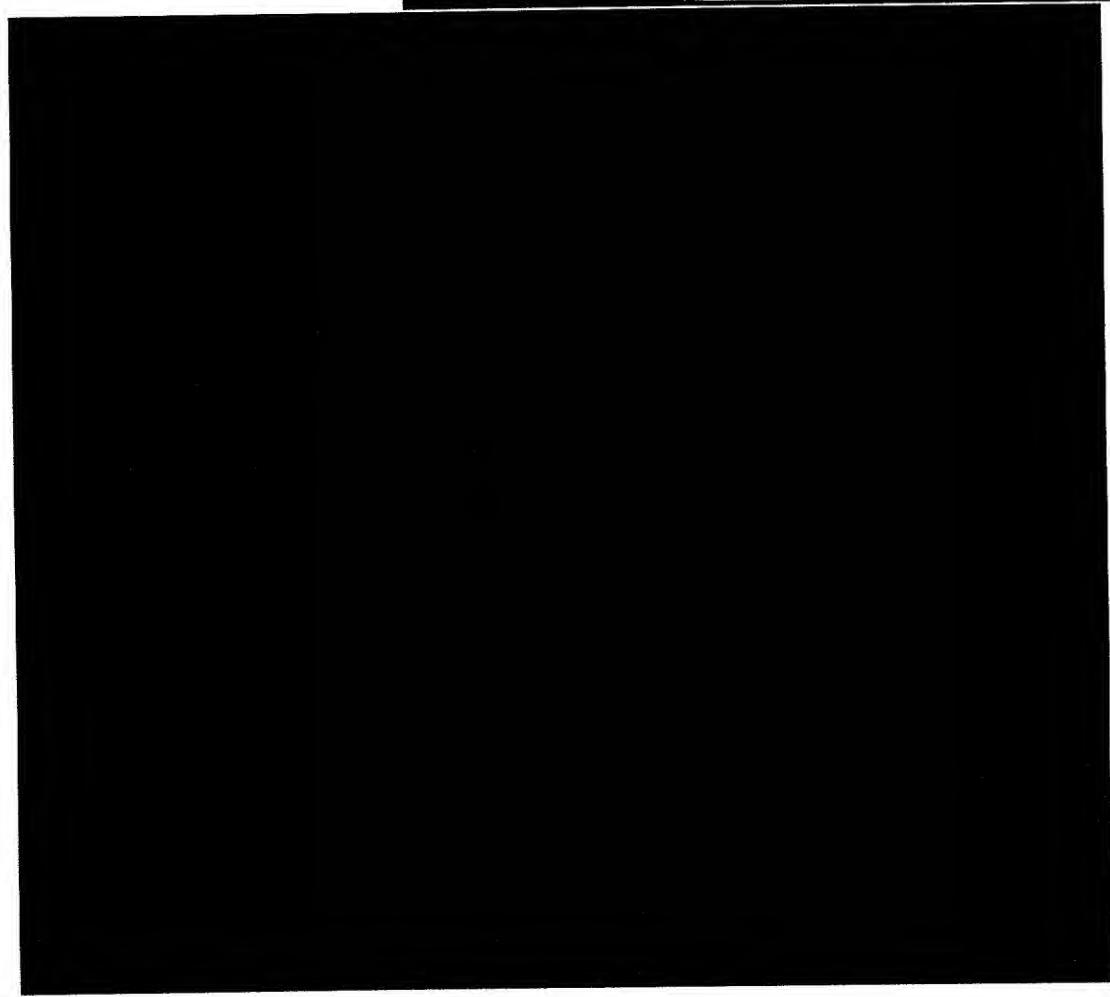


Fig. 1f

Human α GalA-Transduced MSCs in Mouse Muscle: Day-57

10X

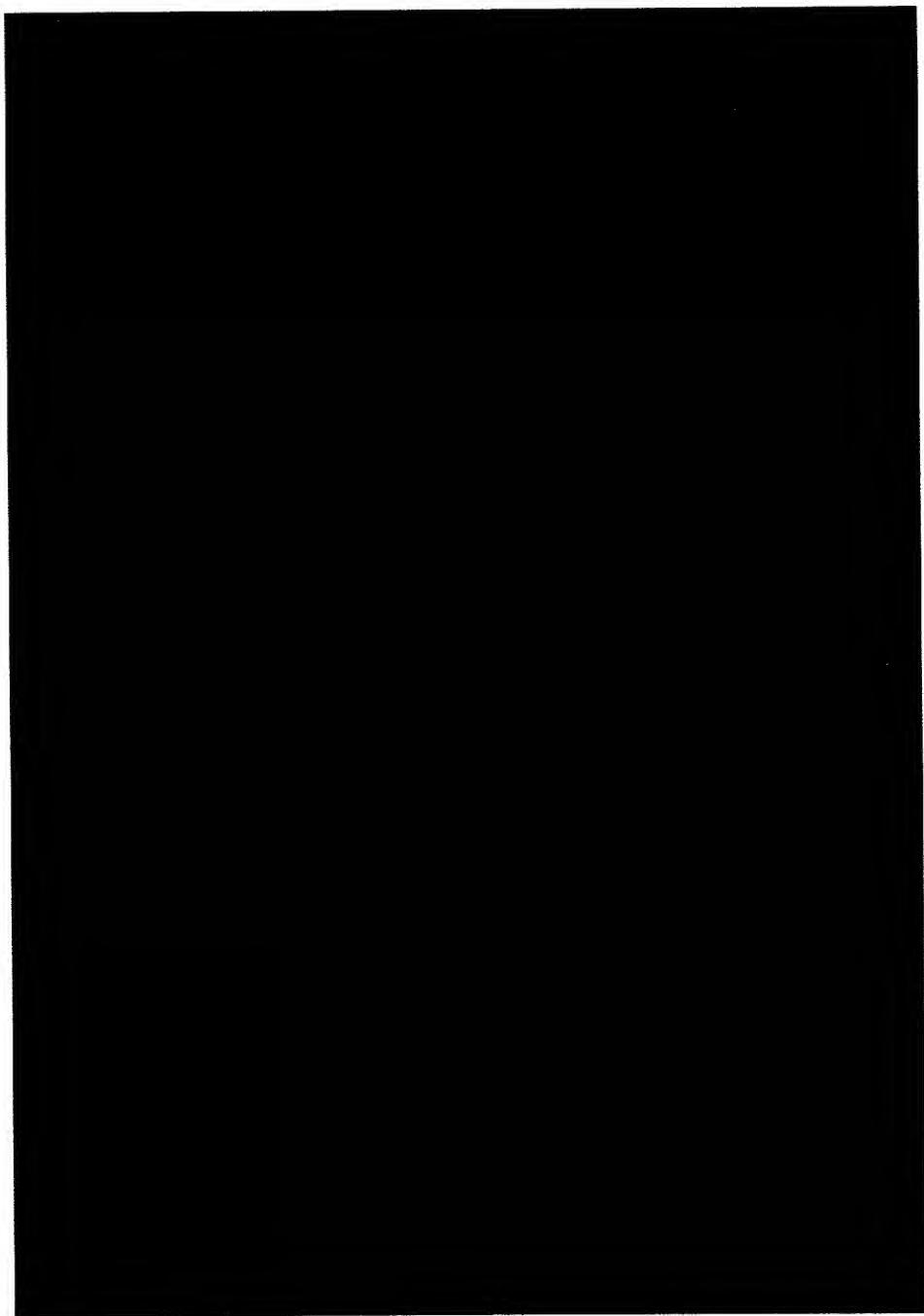


Figure-2
D3: IM injection of Dil-labeled PBMCs

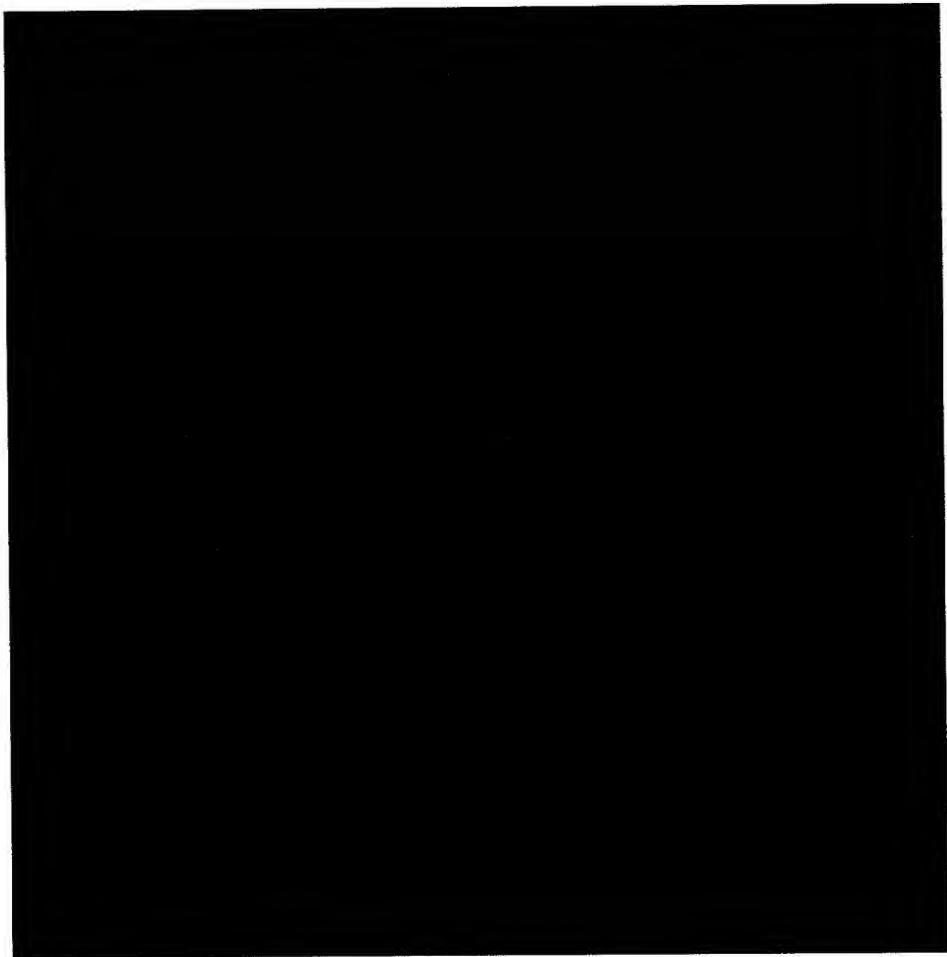
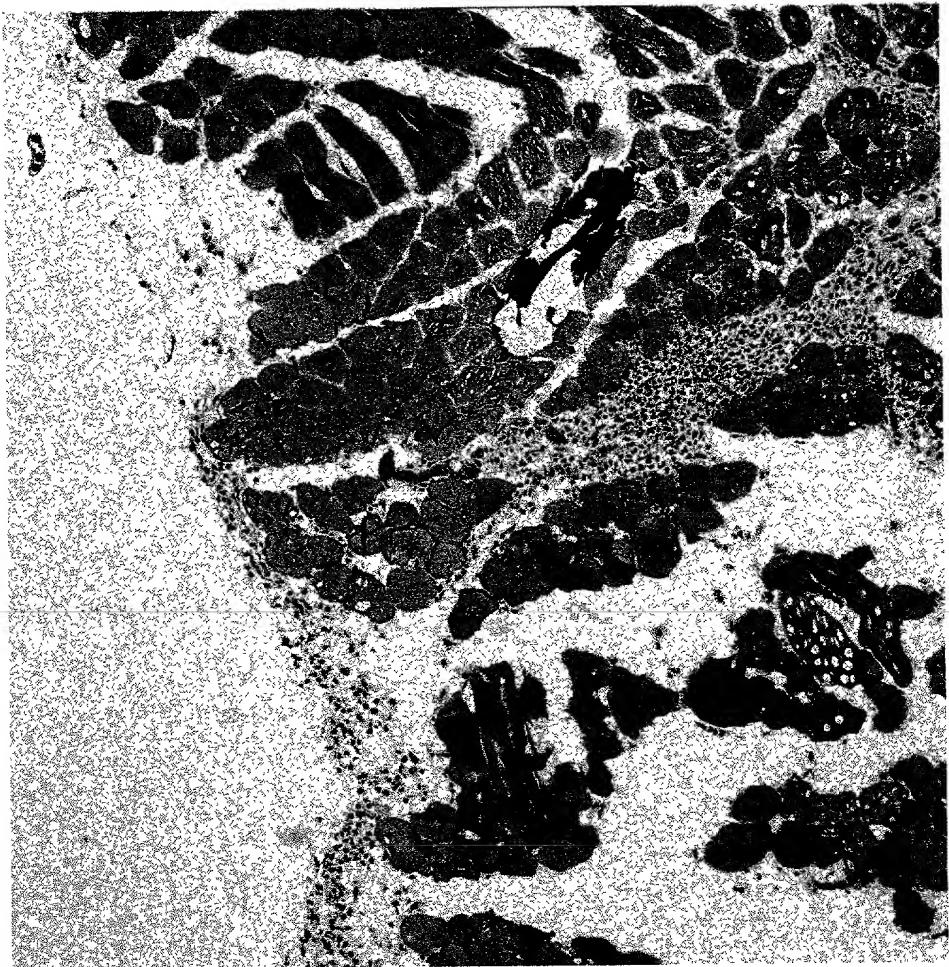
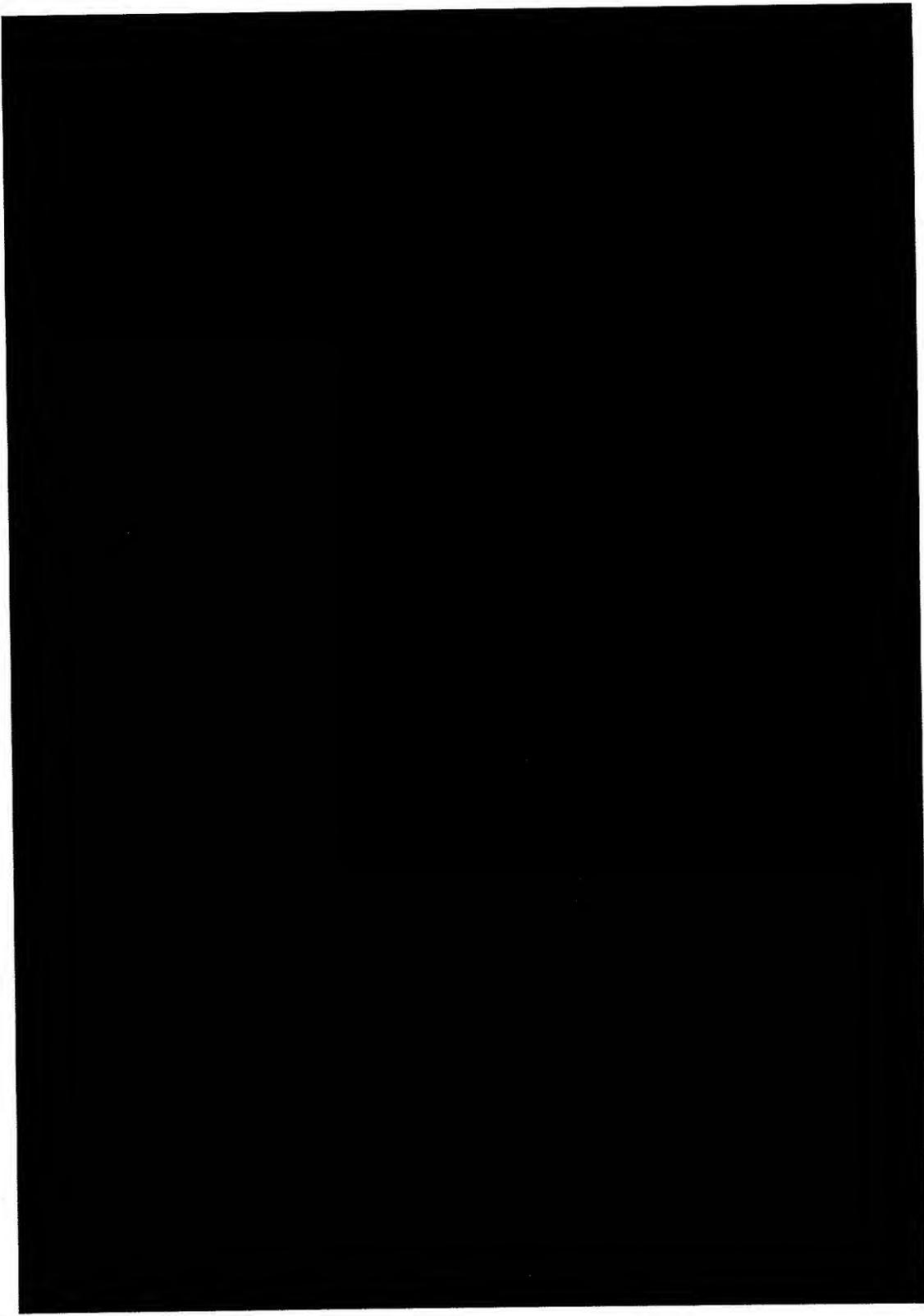


Figure-3
D3 IM injection of saline



Fig. 4 6-week DAPI labeled hMSCs in Mouse Muscle



Red and Blue Fluorescence Channels merged

Fig.5

α GalA Immunostain of Muscle from Mouse
injected IM with α GalA-hMSCs: 14 days

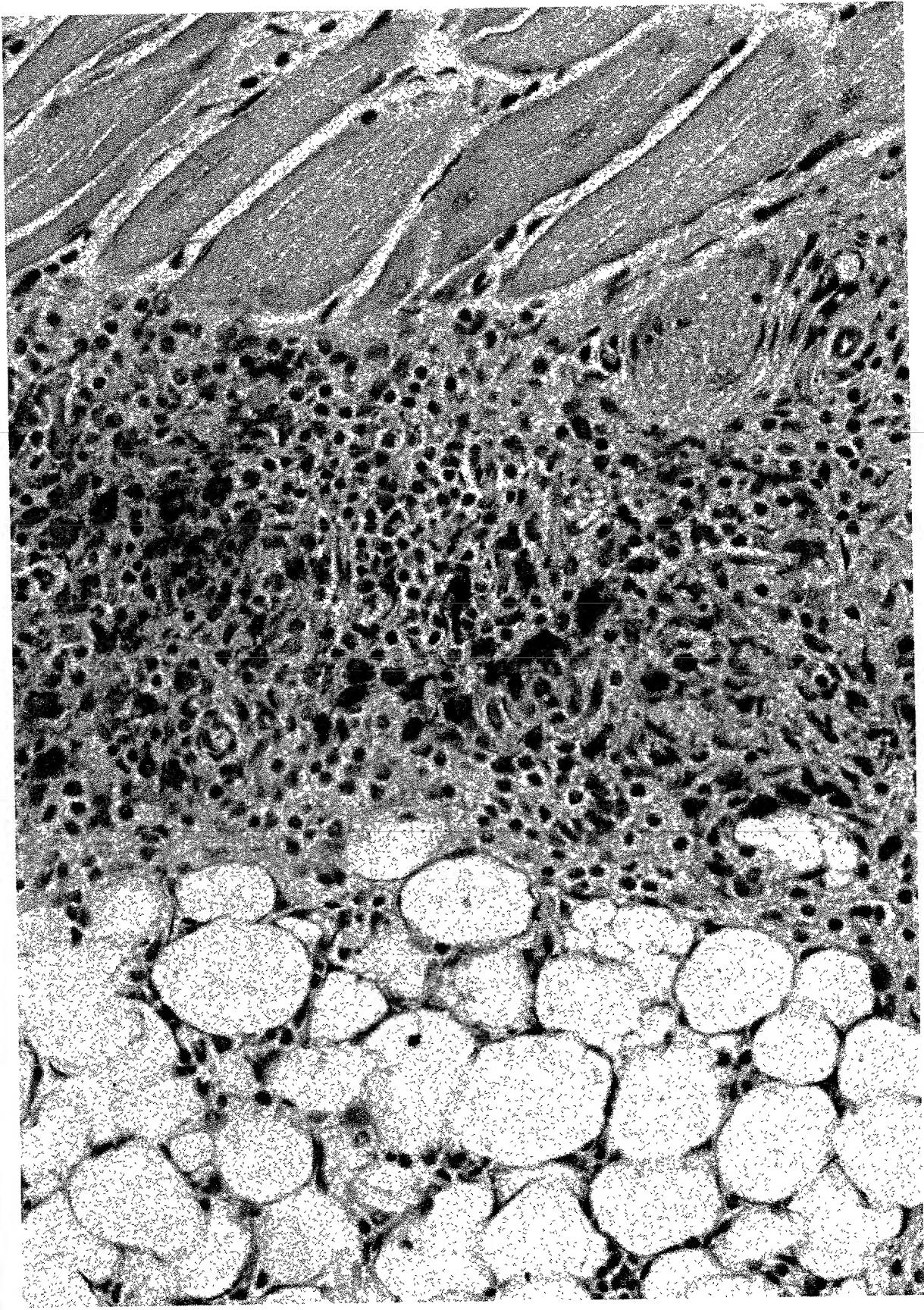


Figure 6

α -GalA activity in Plasma of Mice IM-injected with α -GalA-Transduced hMSCs

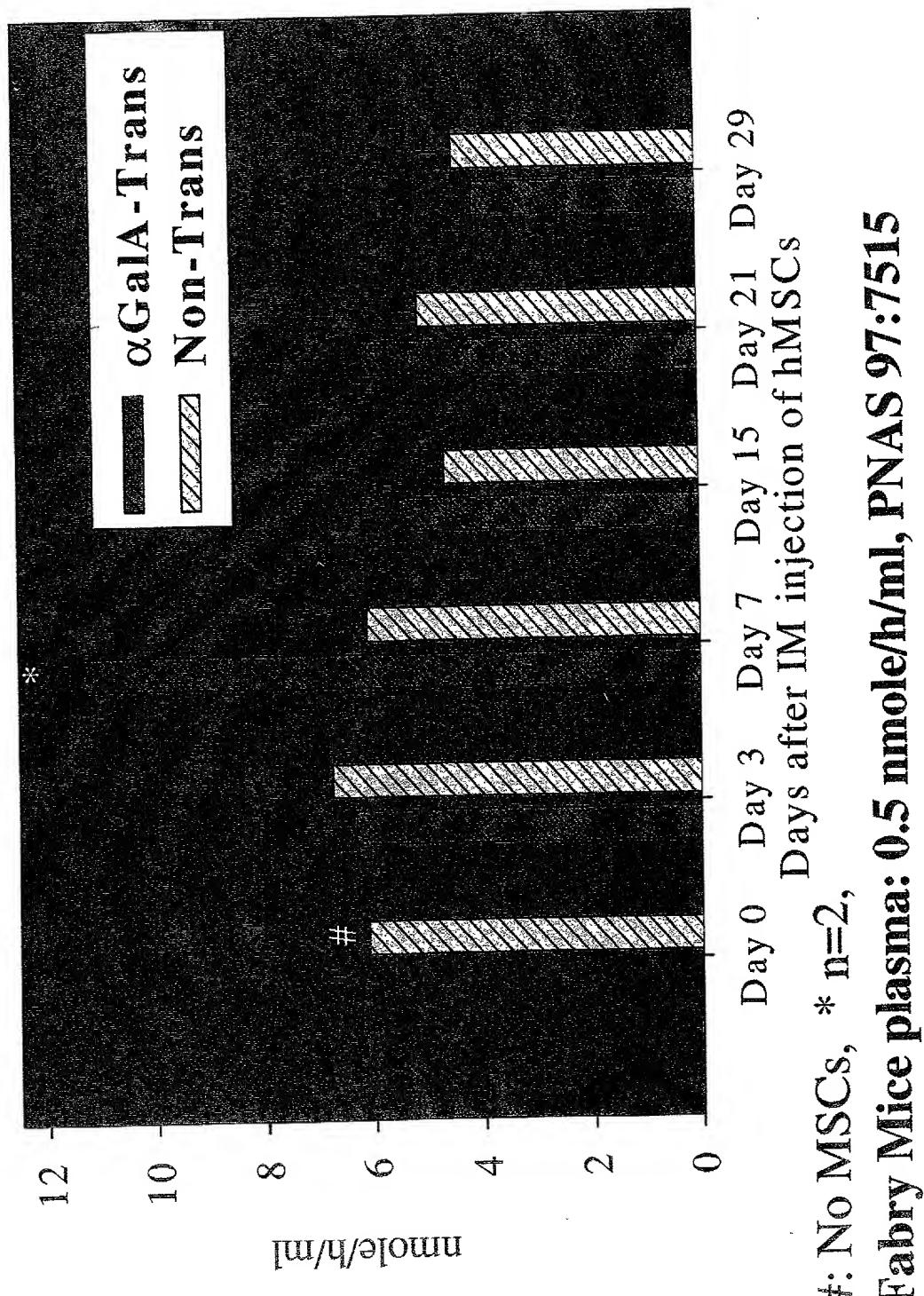
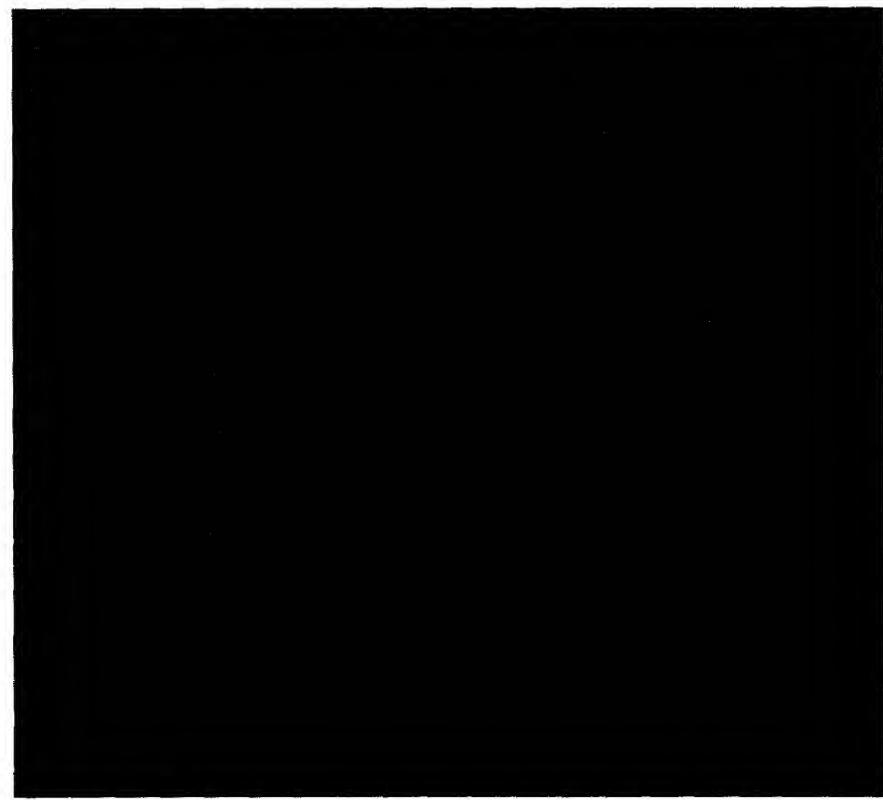


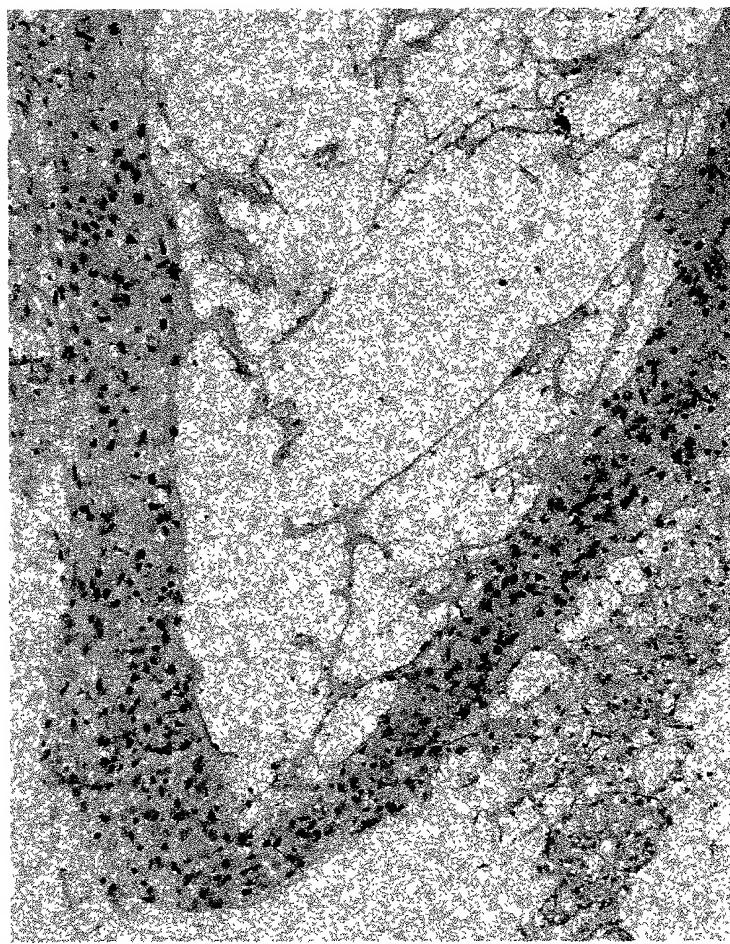
Figure 7a

Human-specific Alu sequence staining of hMSCs injected into Immuno-competent mouse muscle

Dil-positive cells



Alu-sequence positive cells



Human MSCs in Mouse Muscle: 43 days

Figure 7b
H & E

Dil Positive Cells Alu-Positive cells

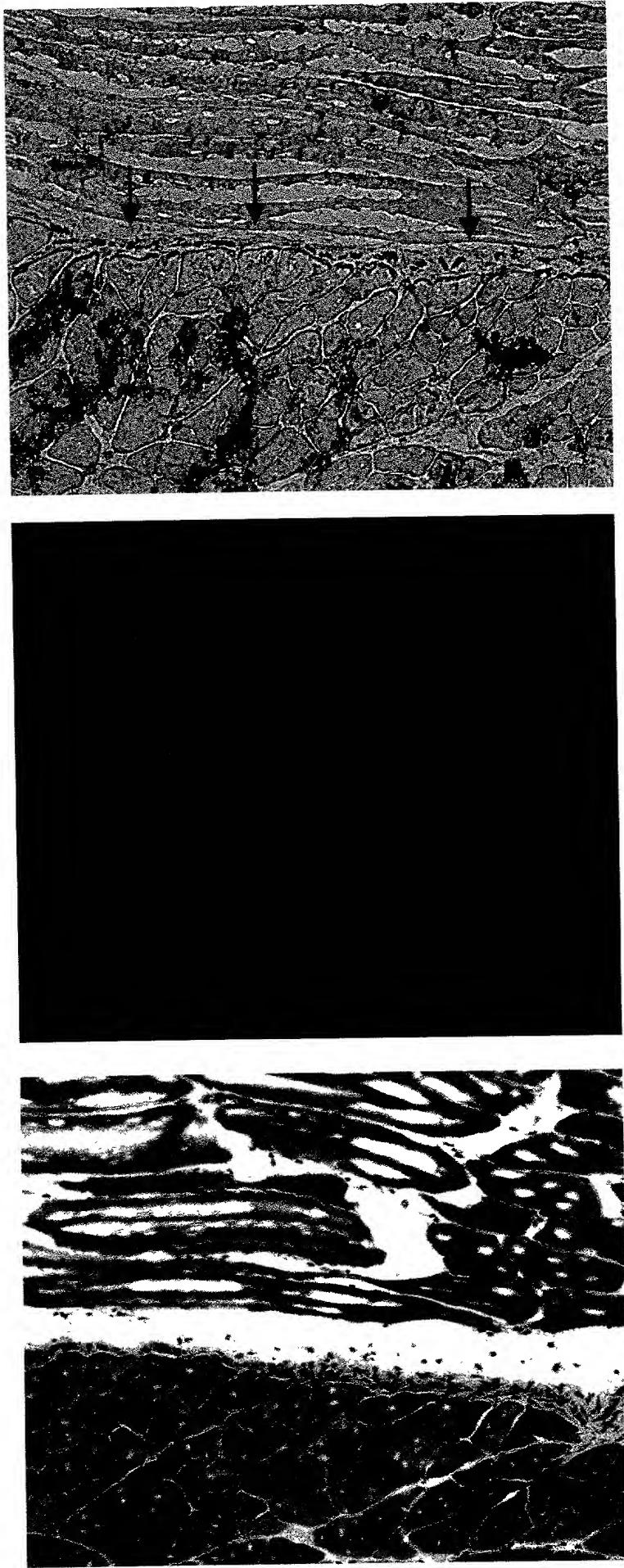
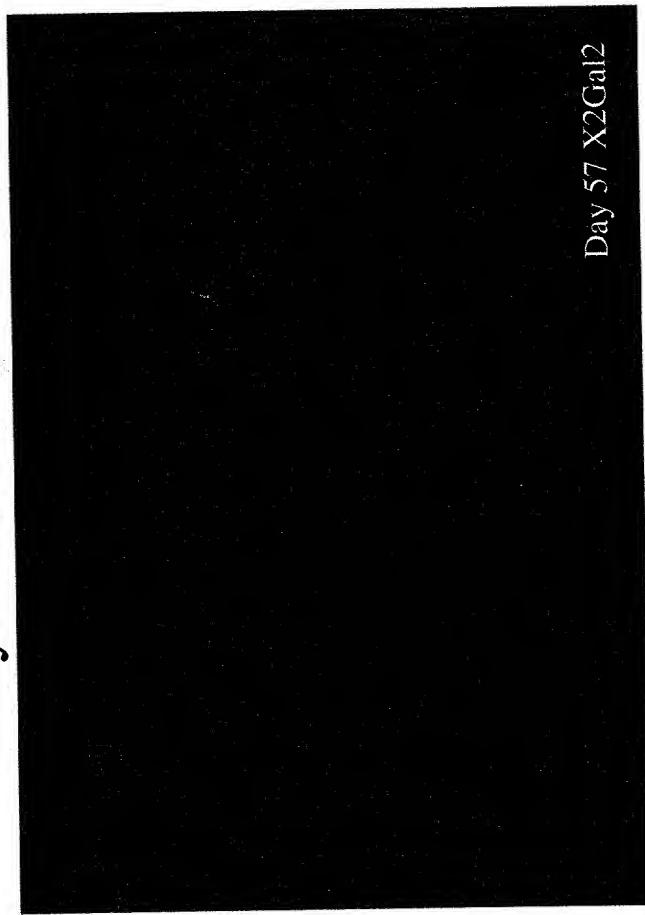


Figure 7c

Day 57 DiI/Alu



Day 57 X2Gal2

-

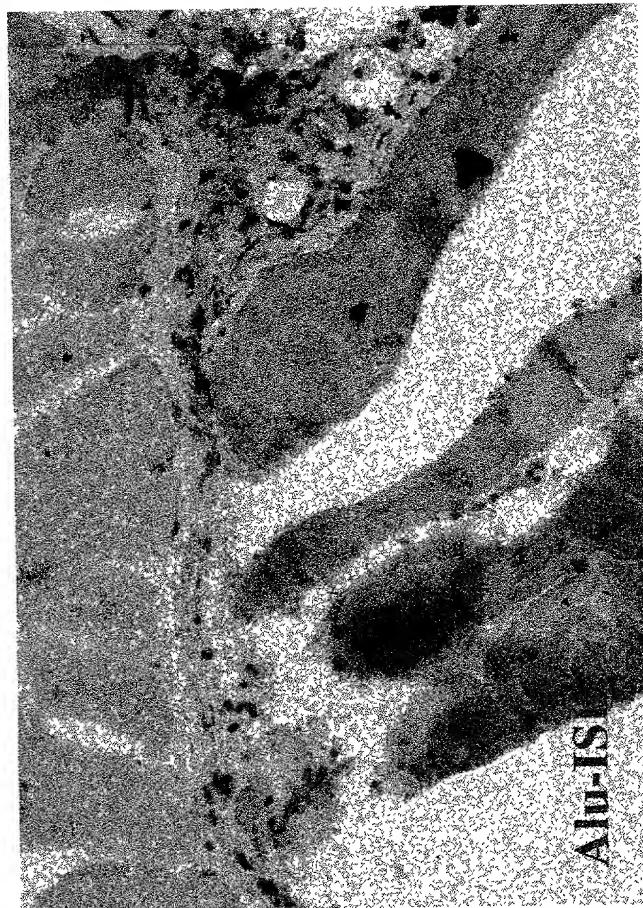
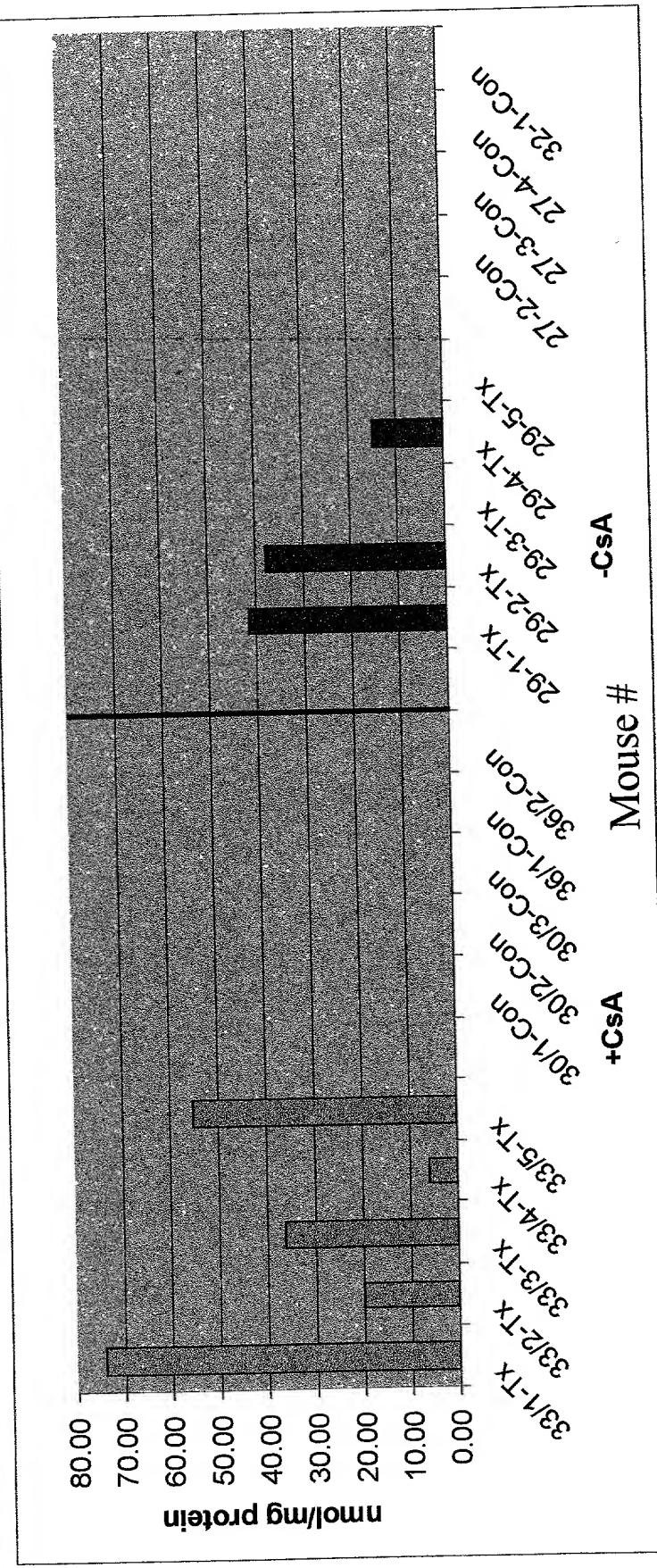


Figure-8a

α GalA Activity in Muscles of Fabry-KO mice:
14 days after iM injection of α GalA-Transduced hMSCs

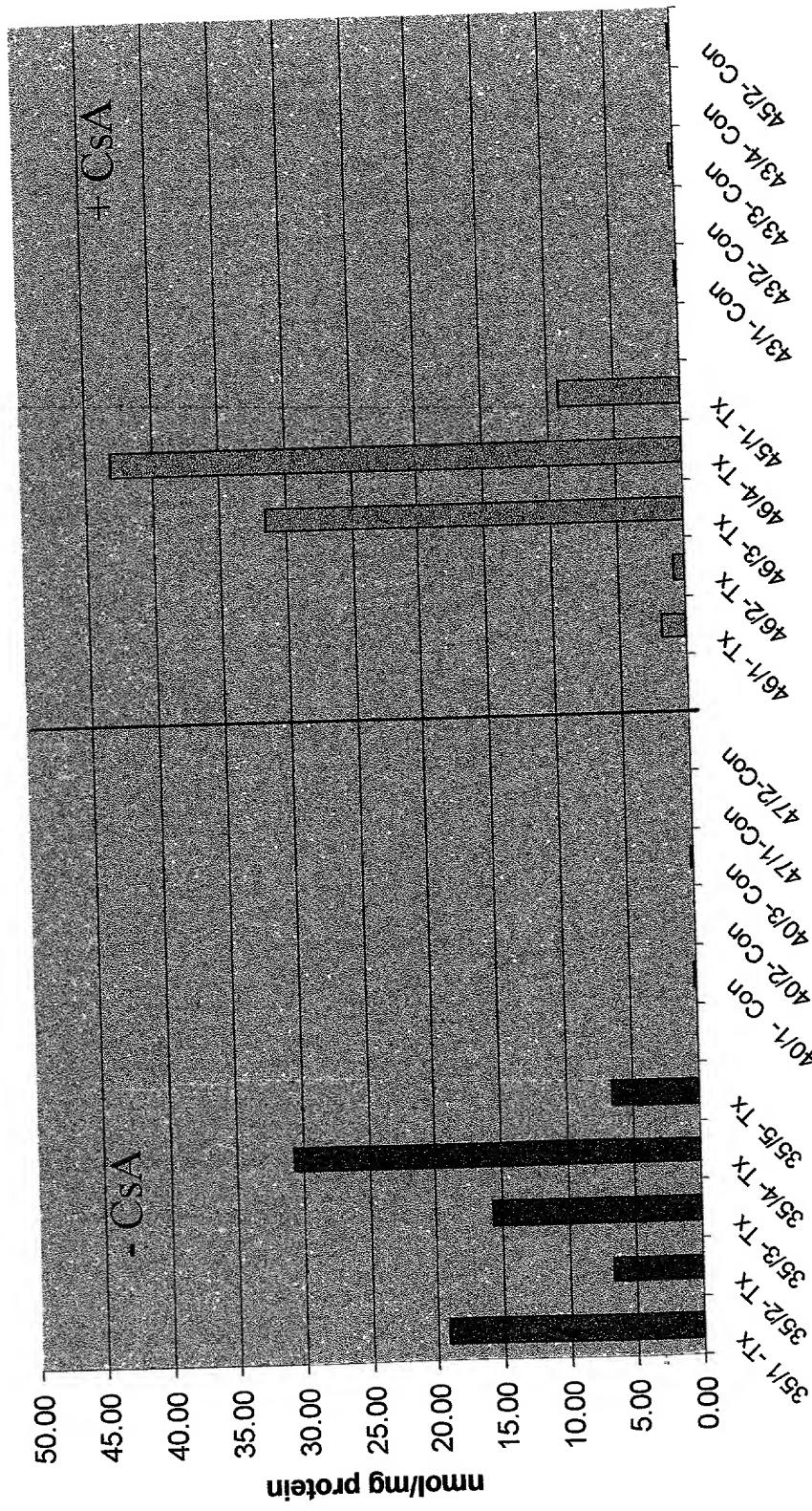


$T_X = \alpha$ GalA Transduced cells injected into muscle

Con = Vehicle alone injected into muscle

Figure-8b

α GaA Activity in Muscles of Fabry-KO mice^a



$T_X = \alpha$ GaLA Transduced cells injected into muscle

C_{01} = Vehicle alone injected into muscle

Figure-**a**
 α GalA Immunostain of Liver from KO Mouse
injected IP with α GalA-hMSCs on Cultispheres :
14 days

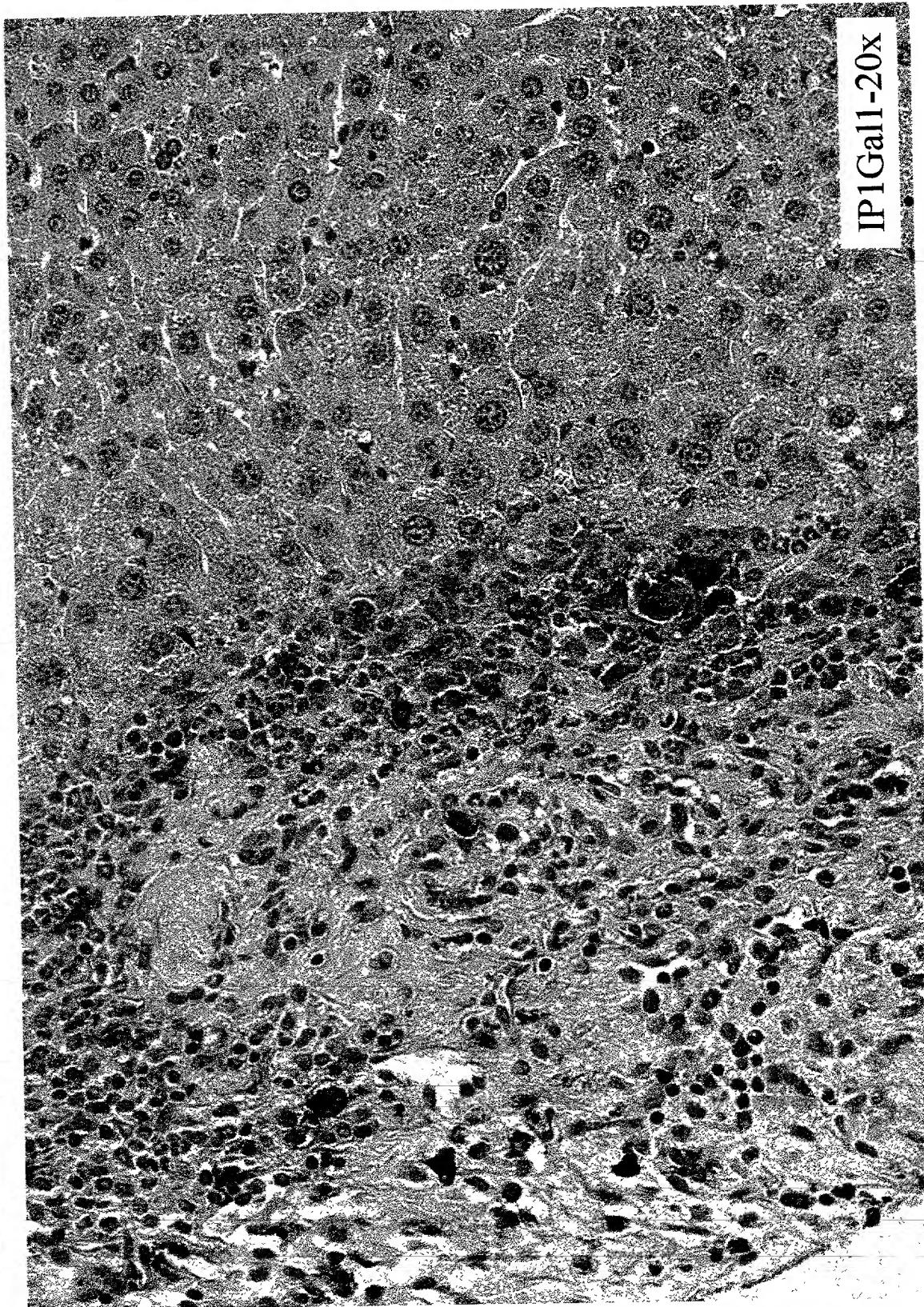


Figure-9b

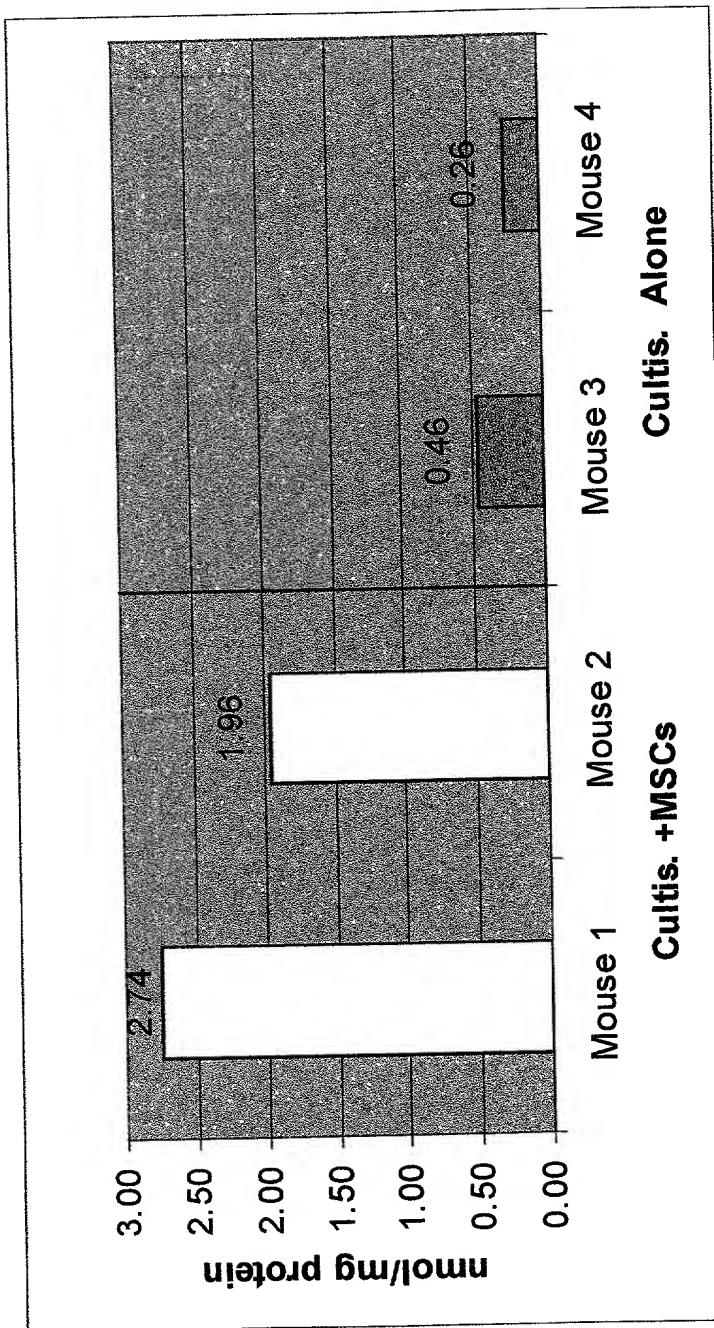
α GalA immunostain of Ewst^{-/-} KO Mouse
injected IP with α GalA-hMSCs on Cultispheres
14 days



Figure 10

Intra-Peritoneal Delivery of α GalA-Transduced hMSCs to Fabry Knock-Out Mice

α GalA Enzyme



Increase in α GalA is 5% of normal mouse liver activity (41 nmole/mg)